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PATIASIDFKRETCVVVYTGYGNREEQNLSDLLSPICEVANNIEQNAQENENESQVSTDE
SENSRSPGNKSDNIKPKSAPWNSFLPPPPPMPGPRLGPGKPGLKFNGPPPPPPPPPPHLL
SCWLPPFPSGPPIIPPPPPICPDSLDDADALGSMLISWYMSGYHTGYYMGFRQNQKEGRC
SHSLN

FIGURE 1

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CGGGGCCCCACGCTGCGCACCCGCGGGTTTGCTATGGCGATGAGCAGCGGCGGCAGTGGT
GGCGGCGTCCCGGAGCAGGAGGATTCCGTGCTGTTCCGGCGCGGCACAGGCCAGAGCGAT
GATTCTGACATTTGGGATGATACAGCACTGATAAAAGCATATGATAAAGCTGTGGCTTCA
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CAACAGTGGAAAGTTGGGGACAAATGTTCTGCCATTTGGTCAGAAGACGGTTGCATTTAC
CCAGCTACCATTGCTTCAATTGATTTTAAGAGAGAAACCTGTGTTGTGGTTTACACTGGA
TATGGAAATAGAGAGGAGCAAAATCTGTCCGATCTACTTTCCCCAATCTGTGAAGTAGCT
AATAATATAGAACAGAATGCTCAAGAGAATGAAAATGAAAGCCAAGTTTCAACAGATGAA
AGTGAGAACTCCAGGTCTCCTGGAAATAAATCAGATAACATCAAGCCCAAATCTGCTCCA
TGGAACCCCTTTCTCCCTCCACCACCCCCCATGCCAGGGCCAAGACTGGGACCAGCAAAG
CCAGGTCTAAAATTCAATGGCCCACCACCGCCACCGCCACCACCACCACCCCACTTACTA
TCATGCTGGCTGCCTCCATTTCCTTCTGGACCACCAATAATTCCCCCACCACCTCCCATA
TGTCCAGATTCTCTTGATGATGCTGATGCTTTGGGAAGTATGTTAATTTCATGGTACATG
AGTGGCTATCATACTGGCTATTATATGGGTTTTAGACAAAATCAAAAAGAAGGAAGGTGC
TCACATTCCTTAAATTAAGGAGAAATGCTGGCATAGAGCAGCACTAAATGACACCACTAA
AGAAACGATCAGACAGATCTGGAATGTGAACCGTTATAGAAGATAACTGGCCTCATTTCT
TCAAAATATCAACTGTTGGGAAAGAAAAAAGGAAGTGGAATGGGTAACTCTTCTTGATTA
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TCTGTAAAAGACTGAGGTGGGGGTGGGAGGCCAGCACGGTGGTGAGGCAGTTGAGAAAAT
TTGAATGTGGATTAGATTTTGAATGATATTGGATAATTATTGGTAATTTTATGGCCTGTG
AGAAGGGTGTTGTAGTTTATAAAAGACTGTCTTAATTTGCATACTTAAGCATTTAGGAAT
GAAGTGTTAGAGTGTCTTAAAATGTTTCAAATGGTTTAACAAAATGTATGTGAGGCGTAT
GTGGCAAAATGTTACAGAATCTAACTGGTGGACATGGCTGTTCATTGTACTGTTTTTTTC
TATCTTCTATATGTTTAAAAGTATATAATAAAAATATTTAATTTTTTTTTTAAAAAAAAAA
AAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAAA
AAAAAAAAAAAAAAAAAAAAAA

FIGURE 2A

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AATTTTTAAATTTTTTGTAGAGACAGGGTCTCATTATGTTGCCCAGGGTGGTGTCAAGCTCCA
GGTCTCAAGTGATCCCCCTACCTCCGCCTCCCAAAGTTGTGGGATTGTAGGCATGAGCCACTG
CAAGAAAACCTTAACTGCAGCCTAATAATTGTTTTCTTTGGGATAACTTTTAAAGTACATTAA
AAGACTATCAACTTAATTTCTGATCATATTTTGTTGAATAAAATAAGTAAAATGTCTTGTGAA
CAAAATGCTTTTTTAACATCCATATAAAGCTATCTATATATAGCTATCTATATCTATATAGCTA
TTTTTTTTTAACTTCCTTTTATTTTCCTTACAG*GGTTTCAGACAAAATCAAAAAGAAGGAAGG
TGCTCACATTCCTTAAATTAAGGA*GTAAGTCTGCCAGCATTATGAAAGTGAATCTTACTTTT
GTAAAACTTTATGGTTTGTGGAAAACAAATGTTTTTGAACAGTTAAAAAGTTCAGATGTTAGA
AAGTTGAAAGCTTAATGTAAAACAATCAATATTAAAGAATTTTGATGCCAAAACTATTAGATA
AAAGGTTAATCTACATCCCTACTAGAATTCTCATACTTAACTGGTTGGTTGTGTGGAAGAAAC
ATACTTTCACAATAAAGAGCTTTAGGATATGATGCCATTTTATATCACTAGTAGGCAGACCAG
CAGACTTTTTTTTTATTGTGATATGGGATAACCTAGGCATACTGCACTGTACACTCTGACATAT
GAAGTGCTCTAGTCAAGTTTAACTGGTGTCCACAGAGGACATGGTTTAACTGGAATTCGTCAA
GCCTCTGGTTCTAATTTCTCATTTGCAG*GAAATGCTGGCATAGAGCAGCACTAAATGACACC
ACTAAAGAAACGATCAGACAGATCTGGAATGTGAAGCGTTATAGAAGATAACTGGCCTCATTT
CTTCAAAATATCAAGTGTTGGGAAAGAAAAAAGGAAGTGGAATGGGTAACTCTTCTTGATTA
AAAGTTATGTAATAACCAAATGCAATGTGAAATATTTTACTGGACTCTTTTGAAAAAC
CATCTGTAAAAGACTGGGGTGGGGGTGGGAGGCCAGCACGGTGGTGAGGCAGTTGAGAAAA
TTTGAATGTGGATTAGATTTTGAATGATATTGGATAATTATTGGTAATTTTATGGCCTGT
GAGAACGGTGTTGTAGTTTATAAAACACTGTCTTAATTTGCATACTTAAGCATTTAGG
AATGAAGTGTTAGAGTGTCTTAAAATGTTTCAAATGGTTTAACAAAATGTATGTGAGGCGT
ATGTGGCAAAATGTTACAGAATCTAACTGGTGGACATGGCTGTTCATTGTACTGTTTTTT
TCTATCTTCTATATGTTTAAAAGTATATAATAAAAATATTTAATTT

FIGURE 2B

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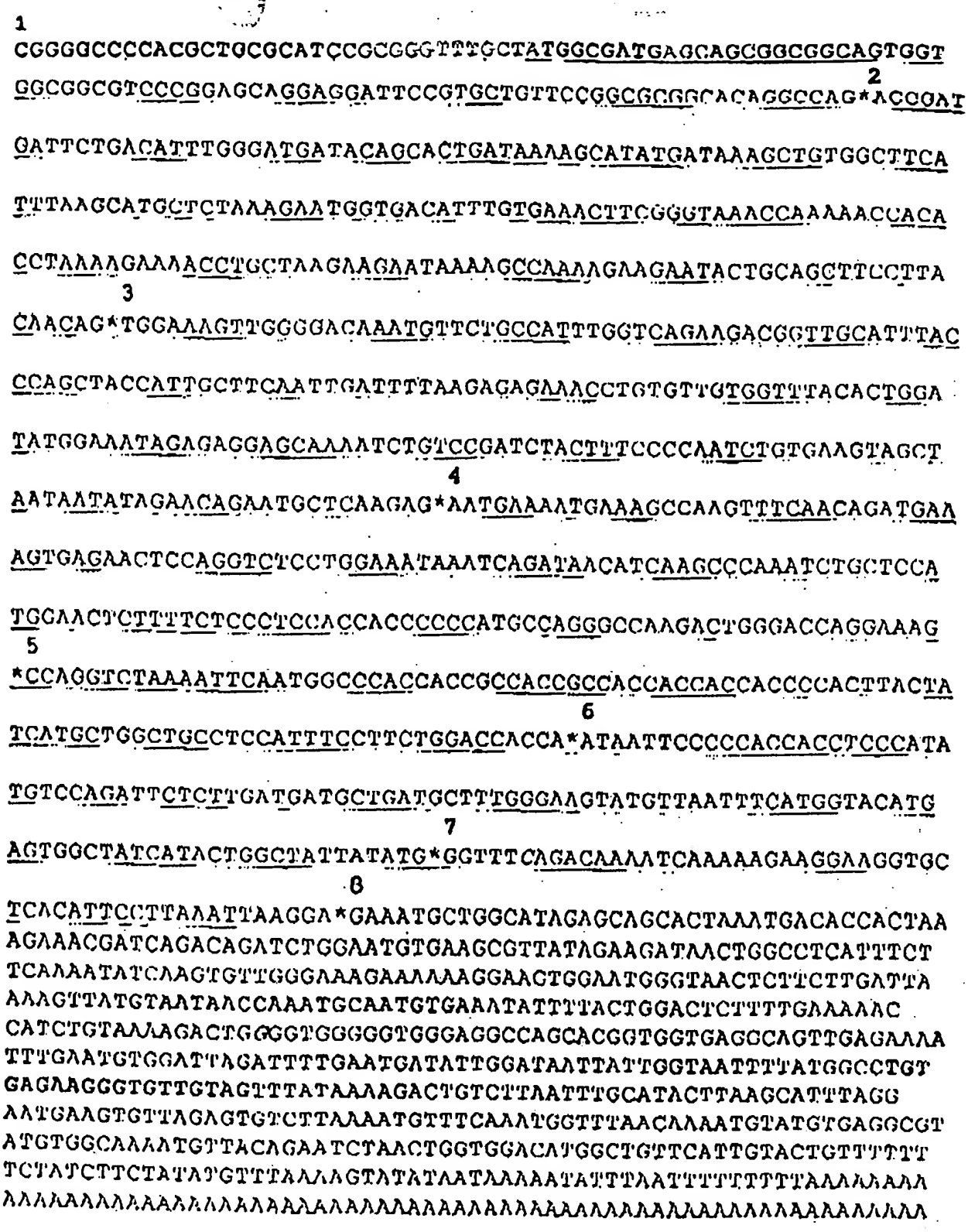

FIGURE 3A

AATTTTTAAATTTTTTGTAGAGACAGGGTCTCATTATGTTGCCCAGGGTGGTGTCAAGCTCCA
GGTCTCAAGTGATCCCCCTACCTCCGCCTCCCAAAGTTGTGGGATTGTAGGCATGAGCCACTG
CAAGAAAACCTTAACTGCAGCCTAATAATTGTTTTCTTTGGGATAACTTTTAAAGTACATTAA
AAGACTATCAACTTAATTTCTGATCATATTTTGTTGAATAAAATAAGTAAAATGTCTTGTGAA
CAAAATGCTTTTTTAACATCCATATAAAGCTATCTATATATAGCTATCTATGTCTATATAGCTA
TTTTTTTTAACTTCCTTTTATTTTCCTTACAG*GGTTTCAGACAAAATCAAAAAGAAGGAAGG
TGCTCACATTCCTTAAATTAAGGA*GTAAGTCTGCCAGCATTATGAAAGTGAATCTTACTTTT
GTAAAACTTTATGGTTTGTGGAAAACAAATGTTTTTGAACAGTTAAAAAAGTTCAGATGTTAAA
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ATACTTTCACAATAAAGAGCTTTAGGATATGATGCCATTTTATATCACTAGTAGGCAGACCAG
CAGACTTTTTTTTTATTGTGATATGGGATAACCTAGGCATACTGCACTGTACACTCTGACATAT
GAAGTGCTCTAGTCAAGTTTAACTGGTGTCCACAGAGGACATGGTTTAACTGGAATTCGTCAA
GCCTCTGGTTCTAATTTCTCATTTGCAG*GAAATGCTGGCATAGAGCAGCACTAAATGACACC
ACTAAACAAACGATCAGACAGATCTGGAATGTGAAGCGTTATAGAAGATAACTGGCCTCATTT
CTTCAAAATATCAAGTGTTGGGAAAGAAAAAAGGAAGTGGAATGGGTAACTCTTCTTGATTA
AAAGTTATGTAATAACCAAATGCAATGTGAAATATTTTACTGGACTCTTTTGAAAAAC
CATCTGTAAAAGACTGGGGTGGGGGTGGGAGGCCAGCACGGTGGTGAGGCAGTTGAGAAAA
TTTGAATGTGGATTAGATTTTGAATGATATTGGATAATTATTGGTAATTTTATGGCCTGT
GAGAAGGGTGTTGTAGTTTATAAAAGACTGTCTTAATTTGCATACTTAAGCATTTAGG
AATGAAGTGTTAGAGTGTCTTAAAATGTTTCAAATGGTTTAACAAAATGTATGTGAGGCGT
ATGTGGCAAAATGTTACAGAATCTAACTGGTGGACATGGCTGTTCATTGTACTGTTTTTT
TCTATCTTCTATATGTTTAAAAGTATATAATAAAATATTTAATTT

FIGURE 3B

C212

ACCTGANCCCAGANGGTCAAGGCTGCAGTGAGACGAGATTGCNCCACTGCCCTCC
ACCCTGGGTGATAAGAGTGGGACCCTGTNTCAAAACATACACACACACACACACA
CACACACACACACACACACACACTCTCTCTCTCTCTCTCTCTCTCTCTCTC
TCTCTCTCTCTCAAAAACACTTGGTCTGTTATTTTTNCGAAATTGTCAGTCAT
AGTTATCTGTTAGACCAAAGCTGNGTAAGNACATTTATTACATTGCCTCCTACAA
CTTCATCAGCTAATGTATTTGCTATATAGCAATTACATATNGGNATATATTATCT
TNAGGGCATGGCCANGTNATAAAACTGTCACTGAGGAAAGGA

C272

CCTCCCACCTNAGCCTCCCCAGTAGCTAGGACTATAGGCGTGCNCCACCAAGCTC
AGCTATTTTTNNTATTTAGTAGAGACGGGGTTTCGGCANGCTTAGGCCTCGTNTC
GAACTCCAGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTGTG
TGTGTAGATATTTATTCCCCCTCCCCCTTGGAAAAGTAAGTAAGCTCCTACTAGG
AATTTAAAACCTGCTTGATCTATATAAAGACAAACAAGGAAAGACAAACATGGGG
GCAGGAAGGAAGGCAGATC

AFM157xd10

TCGAGGTAGATTTGTATTATATCCCATGTACACACACACACACACACACACACAC
ACACACACACACAGACTTAATCTGTTTACAGAAATAAAAGGAATAAAATACCGTT
TCTACTATACACCAAAACTAGCCATCTTGAC

C161

CCCTGAGAAGGCTTCCTCCTGAGTATGCATAAACATTCACAGCTTGCATGCGTGT
GTGTGTGTGTGTGTGTGTGTATGTTTGCTTGCACTGTAAAAACAATTGCAACATC
AACAGAAATAAAAATTAAAGGAATAATTCTCCTCCGACTCTGCCGTTCCATCCAG
TGAAACTCTTCATTCTGGGGTAAAGTTCCTTCAGTTCTTTCATAGATAGGTATAT
ACTTCATAAGTCAAACAATCAGGCTGGGTGCAGTAGCTCATGCCTGTAATCCCAG
CCCTTTGGGAGGCCGAGCTGGGCAGATCGA

C171

TCCACCCGCCTTGGCCTCCCAAAGCNCTGGGATTACAGGCGTGACTGCCGCACCC
AGCTGTAAACTGGNTTNNTAATGGTAGATTTTNAGGTATTAACAATAGATAAAAA
GATACTTTTNGGCATACTGTGTATTGGGATGGGGTTAGAACAGGTGTNCTACCCA
AGACATTTACTTAAAATCGCCCTCGAAATGCTATGTGAGCTGTGTGTGTGTGTGT
GTGTGTGTGTGTATTAAGGAAAAGCATGAAAGTATTTATGCTTGATTTTTTTTT
TNACTCATACCTTCATAGTGGANCAGATACATAGTCTAAATCAAAATGTTTAAAC
TTTTTATGTCACTTGCTGTC

FIGURE 4

19.OCT.95 19:11 No.015 P.08
FAX: 33-1-42660890
GUTMANN-PLASSERAUD

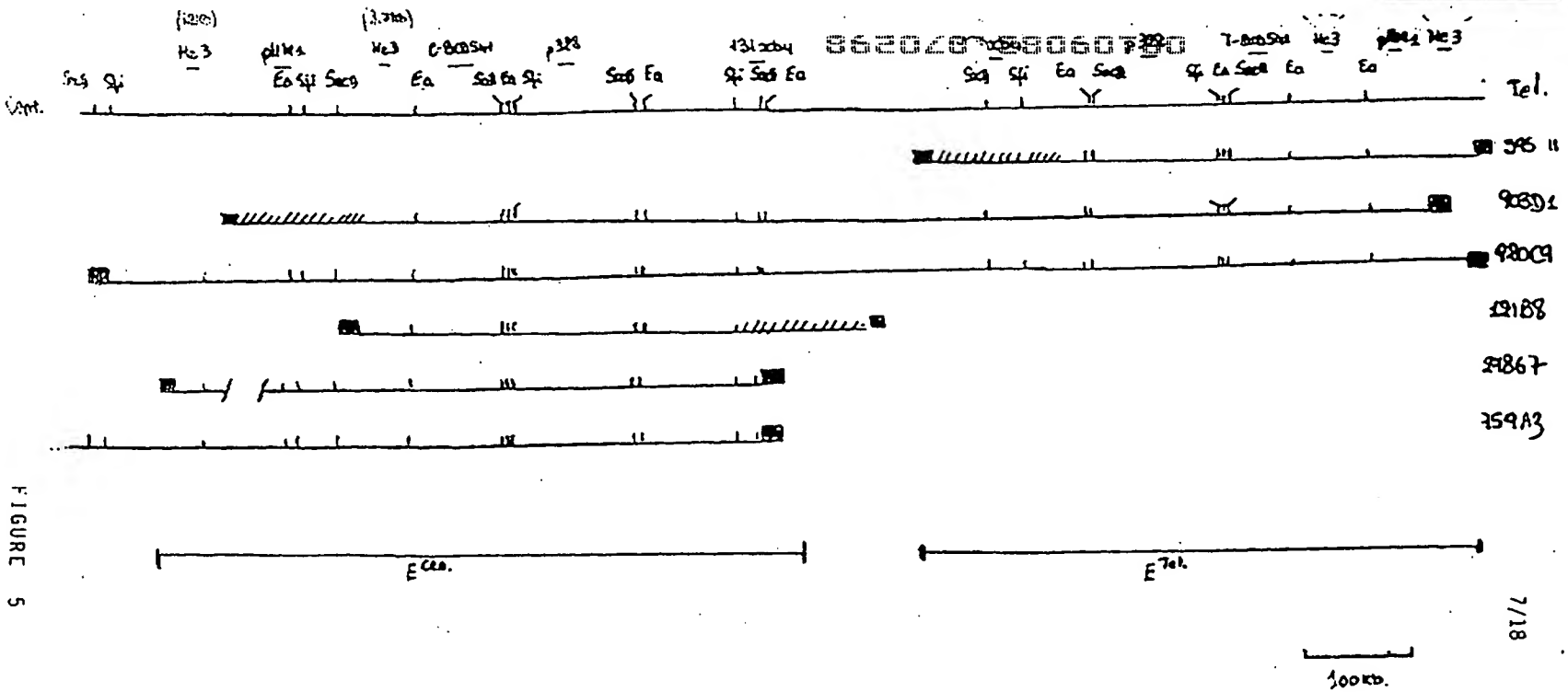
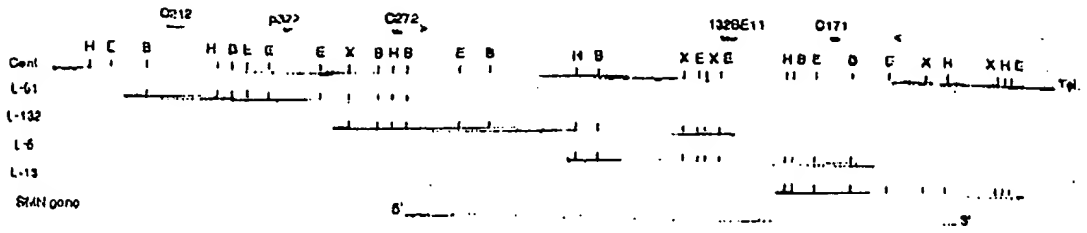


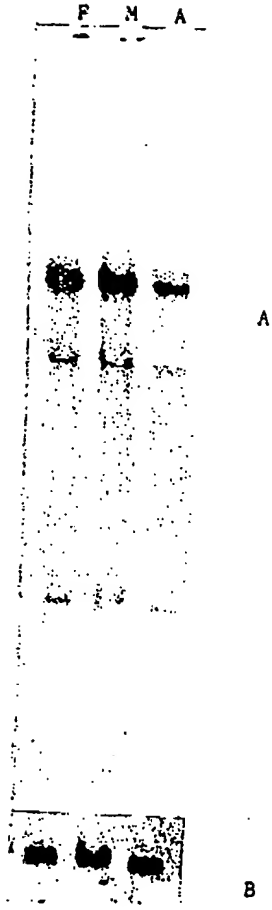
FIGURE 5

Restriction map of the 5q13 region. for Eag I (Ea), Sac II (Sac), Sfi I (Sfi). Numbers under parenthesis indicate the restriction fragment detected by He3; Telomeric element (E^{Tel}), centromeric element ($E^{Cen.}$) Centromere (Cent.), Telomere (Tel.). Probes are indicated above the restriction map. Axes are below the restriction map.



Telomeric element (E^{Tel}) containing the survival motor-neuron gene (SMN gene). Genetic map shows polymorphic markers C212, C272 and C171. Physical map shows location and direction of transcription of SMN gene; phage clones used for assembling physical map. Restriction map for EcoRI(E), XbaI(X), HindIII(H), BglII(B), SacII(S) are shown. Cent. and Tel. indicate centromere and telomere respectively. The position of genomic rearrangements found in SMA patients are also indicated.

FIGURE 6



Gene dosage analysis of the 5q13 region with the 132SE11 plasmid cone in SMA type I patient. Total human DNA from SMA family was digested with HindIII for Southern blotting. Filter was consecutively hybridized with 132SE11 (A) and JK53 probes (B). A significant decrease in 132SE11 band intensity, which indicated the deletion, compared with their parents. F/Father, M/Mother, A/affected

FIGURE 7

MAMSSGGSGGGVPEQEDSVLFRRGTGQSDDSDIWDDTALIKAYDKAVASFKHA
LKNGDICETSGKPKTTPKRKPAKKNKSQKKNTAASLQQWKVGDKCSAIWSEDG
CIYPATIASIDFKRETCVVVYTGYGNREEQNLSDLLSPICEVANNIEQNAQEN
ENESQVSTDESENSRSPGNKSDNIKPKSAPWNSFLPPPPPMPGPRLGPGKPGL
KFNGPPPPPPPPPPPHLLSCWLPPFPSGPPIIPPPPPICPDSLDDADALGSMLI
SWYMSGYHTGYYM

FIGURE 8

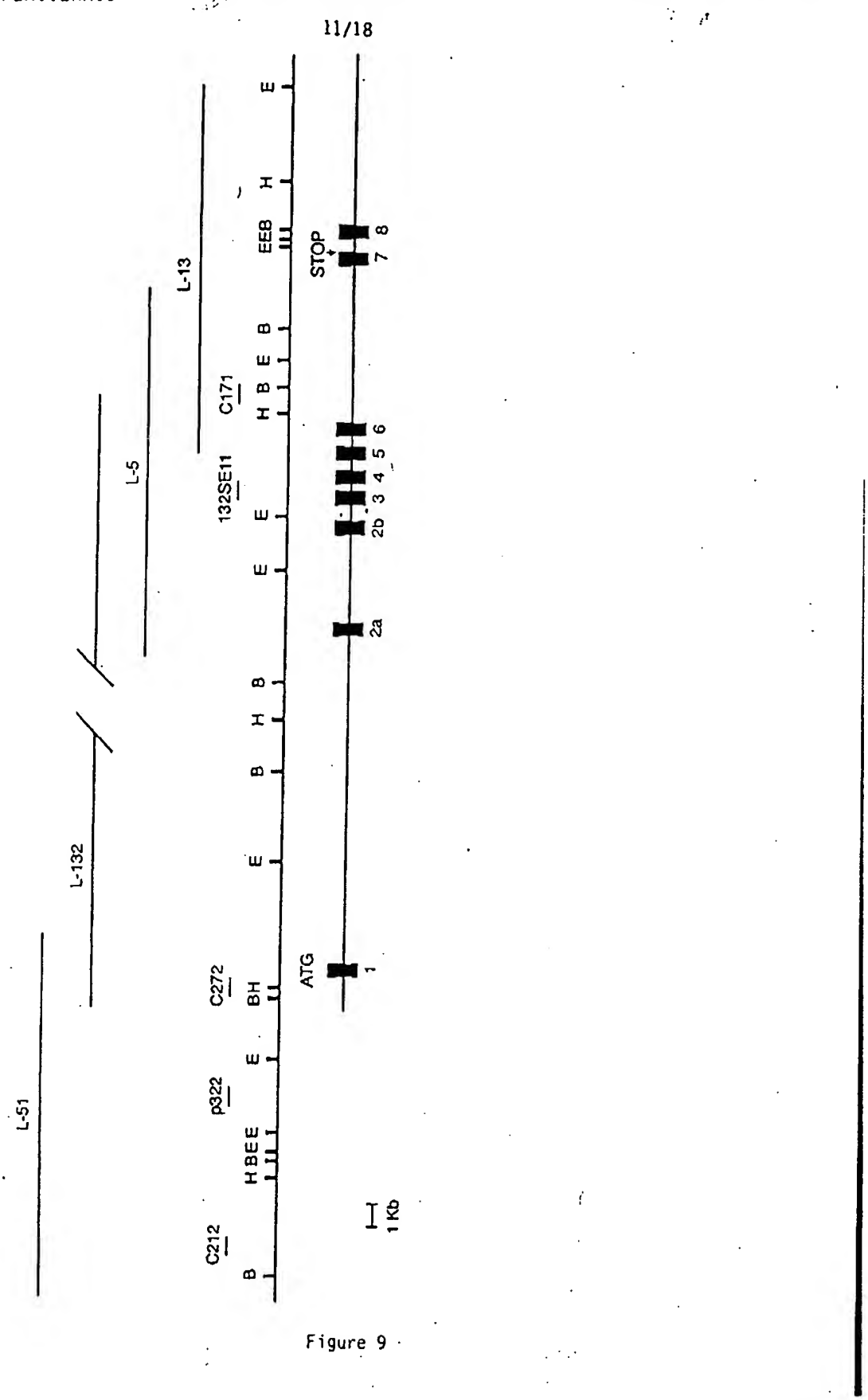

L-51
L-132
L-5
L-13
C212
B
H B E E
p322
E
C272
B H
E
B
H
B
E
132SE11
C171
H B E B
EEB
H
E
ATG
1
2a
2b 3 4 5 6
STOP
7 8
1 Kb

Figure 9

```
1
cctcccgggcaccgtactgttccgctcccagaagccccgggcgccggaagtcgtcactcttaagaagggacg
gggccccacgctgcgcacccgcgggtttgct ATG GCG ATG AGC AGC GGC GGC AGT GGT GGC
                                 M   A   M   S   S   G   G   S   G   G
GGC GTC CCG GAG CAG GAG GAT TCC GTG CTG TTC CGG CGC GGC ACA GGC CAG  gtg
 G   V   P   E   Q   E   D   S   V   L   F   R   R   G   T   G   Q
aggtcgcagccagtgcagtctccctattagcgctctcagcacccttcttccggcccaactctccttccgca
2a
gtgtaattttgttatgtgtggattaagatgactcttggtactaacatacattttctgattaacctatctgn
acatgagttgttttatttcttaccctttccag AGC GAT GAT TCT GAC ATT TGG GAT GAT
                                  S   D   D   S   D   I   W   D   D
ACA GCA CTG ATA AAA GCA TAT GAT AAA GCT GTG GCT TCA TTT AAG  gtatgaaatgc
 T   A   L   I   K   A   Y   D   K   A   V   A   S   F   K
ttgnttagtcgttttcttattttctcgttattcatttggaaaggaattgataacatacgataaagtgttaa
2b
aggtgctttctgaggtgacggagccttgagactagcttatagtagtaactgggttatgtcgtgacttttatt
ctgtgcaccaccctgtaacatgtacatttttattcctattttcgtag CAT GCT CTA AAG AAT GGT
                                                 H   A   L   K   N   G
GAC ATT TGT GAA ACT TCG GGT AAA CCA AAA ACC ACA CCT AAA AGA AAA CCT GCT
 D   I   C   E   T   S   G   K   P   K   T   T   P   K   R   K   P   A
AAG AAG AAT AAA AGC CAA AAG AAG AAT ACT GCA GCT TCC TTA CAA CAG  gttattt
 K   K   N   K   S   Q   K   K   N   T   A   A   S   L   Q   Q
taaaatgttgaggatttaacttcaaaggatgtctcattagtccttatttaatagtgtaaaatgtctttaact
3
gcctgcaggtcgatcaaaacgagatgatagtttgccctcttcaaaagaaatgtgtgcatgtatatatctttg
atttctttgtag TGG AAA GTT GGG GAC AAA TGT TCT GCC ATT TGG TCA GAA GAC
              W   K   V   G   D   K   C   S   A   I   W   S   E   D
GGT TGC ATT TAC CCA GCT ACC ATT GCT TCA ATT GAT TTT AAG AGA GAA ACC TGT
 G   C   I   Y   P   A   T   I   A   S   I   D   F   K   R   E   T   C
GTT GTG GTT TAC ACT GGA TAT GGA AAT AGA GAG GAG CAA AAT CTG TCC GAT CTA
 V   V   V   Y   T   G   Y   G   N   R   E   E   Q   N   L   S   D   L
CTT TCC CCA ATC TGT GAA GTA GCT AAT AAT ATA GAA CAG AAT GCT CAA GAG  gta
 L   S   P   I   C   E   V   A   N   N   I   E   Q   N   A   Q   E
aggatacaaaaaaaaaaaattcaatttctggaagcagagactagatgagaaactgttaaacagtatacaca
4
ccaccgaggcattaatttttcttaatcacacccttataacaaaacctgcatatttttcttttaaag
AAT GAA AAT GAA AGC CAA GTT TCA ACA GAT GAA AGT GAG AAC TCC AGG TCT CCT
 N   E   N   E   S   Q   V   S   T   D   E   S   E   N   S   R   S   P
GGA AAT AAA TCA GAT AAC ATC AAG CCC AAA TCT GCT CCA TGG AAC TCT TTT CTC
 G   N   K   S   D   N   I   K   P   K   S   A   P   W   N   S   F   L
CCT CCA CCA CCC CCC ATG CCA GGG CCA AGA CTG GGA CCA GGA AAG  gtaaaccttct
 P   P   P   P   P   M   P   G   P   R   L   G   P   G   K
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5
caaatattctgggtaattatttttatcctttggtttgagtccttttttattcctatcatattgaaattggt
aagttaatttcctttgaaatattccttatag CCA GGT CTA AAA TTC AAT GGC CCA CCA CCG
                                 P   G   L   K   F   N   G   P   P   P
CCA CCG CCA CCA CCA CCA CCC CAC TTA CTA TCA TGC TGG CTG CCT CCA TTT CCT
 P   P   P   P   P   P   P   H   L   L   S   C   W   L   P   P   F   P
TCT GGA CCA CCA  gtaagtaaaaaagagtataggttagattttgctttcacatacaattgataatta
 S   G   P   P
6
ccagactttacttttgtttactggatataaacaatatctttttctgtctccag ATA ATT CCC CCA
                                                       I   I   P   P
CCA CCT CCC ATA TGT CCA GAT TCT CTT GAT GAT GCT GAT GCT TTG GGA AGT ATG
 P   P   P   I   C   P   D   S   L   D   D   A   D   A   L   G   S   M
TTA ATT TCA TGG TAC ATG AGT GGC TAT CAT ACT GGC TAT TAT ATG  gtaagtaatca
 L   I   S   W   Y   M   S   G   Y   H   T   G   Y   Y   M
ctcagcatcttttcctgacaatttttttgtagttatgtgactttgtttggtaaatttataaaatactacttg
```

Figure 10

7
aactgcagcctaataattgtttctttgggataacttttaaagtacattaaaagactatcaacttaatttct
gatcatattttgttgaataaaataagtaaaatgtcttgtgaaacaaaatgcttttaacatccatataaagc
→ a
tatctatatatagctatctatgtctatatatagtatttttttaacttootttcatttttccttacag GGT
→ T G
TTC AGA CAA AAT CAA AAA GAA GGA AGG TGC TCA CAT TCC TTA AAT taaggagtaag
F R Q N Q K E G R C S H S L N *
tctgccagcattatgaaagtgaatcttactttgtaaactttatggtttgtggaaaacaaatgtttttgaa
→ g
cagttaaaaagttcagatgttaaaaagttgaaaggttaatgtaaaacaatcaatattaaagaattttgatgc
→ g
caaaactattagataaaggttaatctacatccctactagaattctcatacttaactggttggttatgtgga
agaaacatactttcacaataaagagctttaggatatgatgccatttatatcactagtaggcagaccagcag
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aatgggtaactcttcttgattaaaagttatgtaataaccaaatgcaatgtgaaatatttttactggactcttt
→ a
tgaaaaaccatctagtaaaagactgggtgggggtgggaggccagcacggtggtgaggcagttgagaaaatt
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ctgttcattgtactgtttttttctatcttctatatgtttaaaagtatataataaaaatattta

Figure 10 (Continued)

gatctgccttccttcctgcccccatgtttgtctttccttgtttgtctttatatagatcaagcaggttttaaa 72

ttcctagtaggagcttacatttacttttccaaggggggaggggggataaaatatctacacacacacacacac 144

H4TF-1 GH

acaccacactggagttcgagacgaggcctaagcaacatgccgaaaccccgtctctactaaatacaaaaaata 216

DTF-1

gctgagcttggtggcgcacgcctatagtcctagctactgggggaggctgaggtgggaggatcgcttgagccca 288

Sp1

agaagtcgaggctgcagtgagccgagatcgcgccgctgcactccagcctgagcgacagggcgaggctctgtc 360

tcaaaacaaacaaacaaaaaaaaaaaaggaaaggaaatataacacagtgaaatgaaaggattgagagaaatga 432

β-IFN

HiNF-A

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cctcccgggcaccgtactgttccgctcccagaagccccgggcgccggaagtcgtcagtcttaagaagggacg 576

AP-2 E4F1

gggccccacgctgcgcacccgcgggtttgct ATG GCG ATG AGC AGC GGC GGC AGT GGT GGC 637

M A M S S G G S G G

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Figure 11

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gggcgtggtagcaggcc ATG GCG ATG GGC AGT GGC GGA GCG GGC TCC GAG CAG GAA   58
                  Met Ala Met Gly Ser Gly Gly Ala Gly Ser Glu Gln Glu

GAT ACG GTG CTG TTC CGG CGT GGC ACC GGC CAG AGT GAT GAT TCT GAC ATT TGG  112
Asp Thr Val Leu Phe Arg Arg Gly Thr Gly Gln Ser Asp Asp Ser Asp Ile Trp

GAT GAT ACA GCA TTG ATA AAA GCT TAT GAT AAA GCT GTG GCT TCC TTT AAG CAT  166
Asp Asp Thr Ala Leu Ile Lys Ala Tyr Asp Lys Ala Val Ala Ser Phe Lys His

GCT CTA AAG AAC GGT GAC ATT TGT GAA ACT CCA GAT AAG CCA AAA GGC ACA GCC  220
Ala Leu Lys Asn Gly Asp Ile Cys Glu Thr Pro Asp Lys Pro Lys Gly Thr Ala

AGA AGA AAA CCT GCC AAG AAG AAT AAA AGC CAA AAG AAG AAT GCC ACA ACT CCC  274
Arg Arg Lys Pro Ala Lys Lys Asn Lys Ser Gln Lys Lys Asn Ala Thr Thr Pro

TTG AAA CAG TGG AAA GTT GGT GAC AAG TGT TCT GCT GTT TGG TCA GAA GAC GGC  328
Leu Lys Gln Trp Lys Val Gly Asp Lys Cys Ser Ala Val Trp Ser Glu Asp Gly

TGC ATT TAC CCA GCT ACT ATT ACG TCC ATT GAC TTT AAG AGA GAA ACC TGT GTC  382
Cys Ile Tyr Pro Ala Thr Ile Thr Ser Ile Asp Phe Lys Arg Glu Thr Cys Val

GTG GTT TAT ACT GGA TAT GGA AAC AGA GAG GAG CAA AAC TTA TCT GAC CTA CTT  436
Val Val Tyr Thr Gly Tyr Gly Asn Arg Glu Glu Gln Asn Leu Ser Asp Leu Leu

TCC CCG ACC TGT GAA GTA GCT AAT AGT ACA GAA CAG AAC ACT CAG GAG AAT GAA  490
Ser Pro Thr Cys Glu Val Ala Asn Ser Thr Glu Gln Asn Thr Gln Glu Asn Glu

AGT CAA GTT TCC ACA GAC GAC AGT GAA CAC TCC TCC AGA TCG CTC AGA AGT AAA  544
Ser Gln Val Ser Thr Asp Asp Ser Glu His Ser Ser Arg Ser Leu Arg Ser Lys

GCA CAC AGC AAG TCC AAA GCT GCT CCG TGG ACC TCA TTT CTT CCT CCA CCA CCC  598
Ala His Ser Lys Ser Lys Ala Ala Pro Trp Thr Ser Phe Leu Pro Pro Pro Pro

CCA ATG CCA GGT AAC GGA TTA GGA CCA GGA AAG CCA GGT CTA AAA TTC AAC GGC  652
Pro Met Pro Gly Asn Gly Leu Gly Pro Gly Lys Pro Gly Leu Lys Phe Asn Gly

CCG CCG CCG CCG CCT CCA CTA CCA CCT CCC CCA TTC CTG CCG TGC TGG ATG CCC  706
Pro Pro Pro Pro Pro Pro Leu Pro Pro Pro Pro Phe Leu Pro Cys Trp Met Pro

CCG TTC CCT TCA GGA CCA CCA ATA ATC CCG CCA CCC CCT CCC ATC TCT CCC GAC  760
Pro Phe Pro Ser Gly Pro Pro Ile Ile Pro Pro Pro Pro Pro Ile Ser Pro Asp

TGT CTG GAT GAC ACT GAT GCC CTT GGC AGT ATG CTA ATC TCT TGG TAC ATG AGT  814
Cys Leu Asp Asp Thr Asp Ala Leu Gly Ser Met Leu Ile Ser Trp Tyr Met Ser

GGC TAC CAC ACT GGC TAT TAT ATG GGT TTC AGA CAA AAT AAA AAA GAA GGA AAG  868
Gly Tyr His Thr Gly Tyr Tyr Met Gly Phe Arg Gln Asn Lys Lys Glu Gly Lys

TGC TCA CAT ACA AAT TGA
Cys Ser His Thr Asn  *                                                   887
```

Figure 12

```
 20        30        40        50        60        70        80
GSGGGVPEQEDSVLFRRGTGQSDDSDIWDDTALIKAYDKAVASFKHALKNGDICETSGKPKTTPKRKPAK
====   ====-=========================================== ===-= -=====
GSGGAGSEQEDTVLFRRGTGQSDDSDIWDDTALIKAYDKAVASFKHALKNGDICETPDKPKGTARRKPAK
          20        30        40        50        60        70
 90       100       110       120       130       140       150
KNKSQKKNTAASLQQWKVGDKCSAIWSEDGCIYPATIASIDFKRETCVVVYTGYGNREEQNLSDLLSPIC
========--- = ============-===========-========================== =
KNKSQKKNATTPLKQWKVGDKCSAVWSEDGCIYPATITSIDFKRETCVVVYTGYGNREEQNLSDLLSPTC
          90       100       110       120       130       140
160       170       180       190       200       210       220
EVANNIEQNAQENFNESQVSTDESENSRSPGNKSDNIKPKSAPWNSFLPPPPPMPGPRLGPGKPGLKFNG
====  ===-====--=======-== =             = =-=== ==========  ===========
EVANSTEQNTQENE--SQVSTDDSEHSSRSLRSKAHSKSRAAPWTSFLPPPPPMPGSGLGPGKPGLKFNG
        160        170       180       190       200       210
230       240       250       260       270       280       290
PPPPPPPPPPHLLSCWLPPFPSGPPIIPPPPPICPDSLDDADALGSMLISWYMSGYHTGYYMGFRQNQKE
====== === = ==-=============== == ===-========================== ==
PPPPPPLPPPPFLPCWMPPFPSGPPIIPPPPPISPDCLDDTDALGSMLISWYMSGYHTGYYMGFRQNKKE
220       230       240       250       260       270       280
300
GRCSHSL
=-===-
GKCSHTN
290
```

Figure 13

0520702 : 2006076D

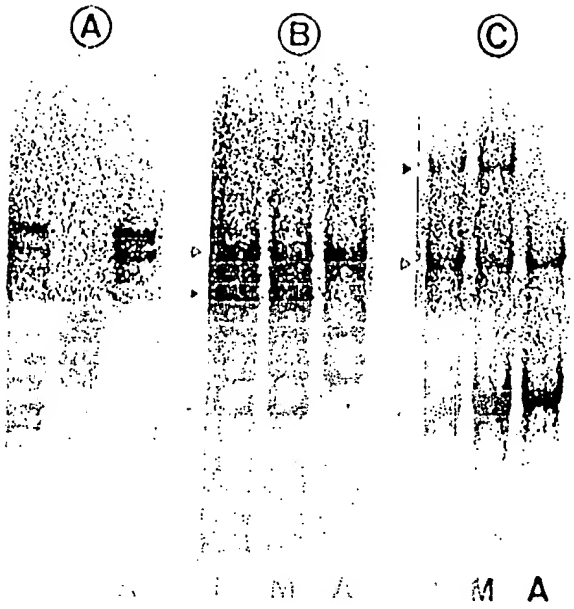

Figure 14

SSCP Analysis

▼ SMN

▽ C-BCD541

421B8 YAC

595C11 YAC

HUMAN 1 CONTROL

HUMAN 2 CONTROL

HUMAN 3 CONTROL

HUMAN 4 SMA

Figure 1